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atous Tumors, with the Report
of a Case and Presentation
of the Specimen.

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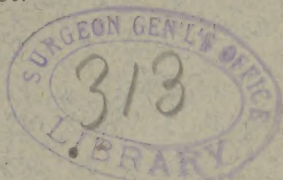
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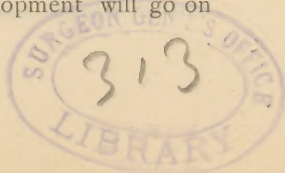
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FIBRO- OR SPINDLE-CELLED SARCOMATOUS TUMORS, WITH THE REPORT OF A CASE AND PRESENTATION OF THE SPECIMEN.

Various names have been employed by modern authors to designate these tumors, and such appellations are commonly expressive of their structural composition or pathological peculiarities. We accordingly find Mr. Paget speaking of them as "recurrent fibroids;" Lebert, as "fibro-plastic tumors;" Müller, "fasciculated carcinoma;" Gluge, "albuminous sarcoma;" Cornil and Ranvier, "fasciculated sarcoma;" Follin, "plasmona;" Bennett, "fibro-nucleated tumors;" Lancereaux, "spindle-celled sarcomatous tumors;" while the same morbid condition is discussed by Billroth under the head of "spindle-celled sarcoma or fibro-sarcoma." These tumors vary greatly in size and may not be larger than a filbert in one instance, while, in other cases, the dimensions occasionally exceed those of the adult human head.

These neoplasms were first described by Lebert under the name of "fibro-plastic tumors." They are very common and may be developed in various regions of the body. They are most frequently found in the skin and subcutaneous structures, but likewise frequently occur in the deep seated connective tissue, aponeuroses, mammary gland, testicles, and also, occasionally, in the muscular tissue. The disease may also originate in the periosteum and by some authors it is thought possible for it to find a starting point even in the endosteum of the long bones. These tumors are never uniform in their external appearance, inasmuch as the form of the growth is commonly dependent on the surrounding tissues, and it is a well known fact that development will go on



most rapidly in the direction in which it meets the least resistance. These growths are generally rounded or lobulated and firmly attached to the surrounding tissues, and even a casual examination after their removal, shows them to be firm, tough, and irregular. A transverse section of the tumor having been made, brings into view a pale red, pale pink, or clear grayish-white tissue, and the surface has a streaked appearance as if from the presence of fibrous bands and bundles of fibrous tissue variously arranged, sometimes straight, sometimes curved, at other times parallel or even crossing each other, and occasionally found in concentric layers, similar to that which is observed on the section of uterine myoma. There is not seen, on the freshly cut surface of these tumors, any juice, or at least only a very small quantity of a transparent fluid, but in those tumors which have been left exposed to the air after their removal, until decomposition has set in, it will be observed that if incisions are now made in them, there soon appears on the cut surfaces a whitish, milk-colored fluid, which, however, is less in quantity than that commonly exuded under the same circumstances when some of the other varieties of sarcoma are incised during the stage of decomposition.

The cell formation of the spindle-celled sarcoma is copied very accurately after the spindle-celled tissue of recent cicatrices, as the microscopical examination of these tumors shows that they are made up of a short and narrow spindle-cell with an elongated, roundish nucleus, with or without nucleoli, enlarged in the middle and terminated in two fine points; it is also this peculiar form which has caused them to be compared with the spindle; this peculiarity supplying the explanation why the name of spindle-cell has been given to these elements and the name, spindle-celled sarcoma applied to these morbid growths. The spindle-cells vary greatly in size, and this fact has caused Rindfleisch to designate the spindle-celled

sarcomatous growths as the "small spindle-celled sarcoma," and the "large spindle-celled sarcoma." This distinction possesses but little value since all the intermediate sizes are found in our pathological examinations; nevertheless, it may be well to remember that the small spindle-cells characterize a tumor which is more dangerous to life—more prone to return when it has been removed. It therefore seems that the large spindle-cells represent a more favorable condition of the elements—the prognosis is less grave, and consequently from the clinical stand-point, it is not a matter of entire indifference whether the cells are large or small. Besides these spindle-cells, there is frequently found a certain number of round cells, similar to those observed in cases of encephaloid sarcoma. The latter are sometimes very limited in number, whilst, on the contrary, they may be very numerous. In these instances the surgeon is dealing with a mixed form of sarcoma, intermediate between the encephaloid and the spindle-celled.

The arrangement of the fibrous bundles in the spindle-celled sarcoma is very simple, the fusiform cells being placed side by side in such a manner that the enlarged portion of one cell is in contact with the slender part of its neighbor, and these are firmly united by an intercellular substance. The uniting, or intercellular substance is hyaline, more firm and less abundant than in cases of encephaloid sarcoma. The vessels in these tumors possess few peculiarities; the larger are observed running in the same direction as the larger fasciculæ, but the smaller do not conform to this rule. The tissue of the spindle-celled sarcoma is more dense than that of the encephaloid, and consequently interstitial hæmorrhage is more rare. In the encephaloid variety it has been observed that fatty granular changes, mucous transformation, etc., are frequent occurrences which are not seen in cases of the spindle-celled sarcoma.

It is highly important that sarcoma should be care-

fully studied in connection with the tumors of the female breast, and although these sarcomatous growths may be either encephaloid or spindle-celled, it is, nevertheless, highly important, from both a diagnostic and prognostic standpoint, that the surgeon should be able to make a differential diagnosis of sarcoma and carcinoma of the breast. This difference consists of well marked pathological characteristics, which I shall attempt to portray in the following diagram.

DIFFERENTIAL DIAGNOSIS OF SARCOMA AND CARCINOMA OF THE BREAST.

SARCOMA.

1. Commonly develops very slowly, especially at first; may remain stationary for years.

2. Rough, lobulated, or lumpy; lump may be as large as a hen's egg; tumor finally attaining great size and becoming pediculated.

3. Skin involved after a long interval; morbid growth approaches the integument, which is gradually thinned as by an abscess, and also frequently marked with large veins.

4. Nipple does not retract and is not often changed in appearance.

5. Ulceration occurs after the lapse of a long period; skin gives way, owing to pressure on its internal surface by the lumps which belong to the morbid growth, but the ulcerated border of the integument is thin, loose and not adherent to the tumor.

CARCINOMA.

1. Commonly develops very rapidly and may terminate fatally within a year.

2. Slightly roughened; no large lobules; tumor usually small and flattened on the chest.

3. Skin becomes quickly attached to the morbid growth, is retracted, drawn in, thus giving rise to the appearance of a quilted cover; large veins not seen, but in their stead there may be observed white lines, sometimes called lymphatic varices.

4. Nipple retracts and its end seems to be absorbed.

5. Ulceration occurs at an early date; skin is invaded by the morbid growth, and destroyed; border is thickened, hardened and adherent to the tumor.

6. Consistence of the tumor varies in the different stages of the disease; first hard, later soft spots may be found, and even liquid parts from the cysts within it.

7. The mammary gland remains distinct from the tumor; consequently it is not destroyed, but simply flattened and atrophied.

8. Sarcoma does not become adherent to the deep-seated parts.

9. Does not involve the lymphatic system in the early stage of the disease, and rarely, even in the late.

10. The morbid growth returns in the majority of cases, commonly in the same organ, and these relapses indicate a finally fatal termination of the disease.

11. The general health of the patient often remains quite satisfactory, even after the tumor has been removed several times.

12. The progress of the disease is rarely attended with much pain.

6. Consistence of the tumor never varies in the different stages. Generally firm.

7. This morbid growth, from the first, fuses with the mammary gland and soon destroys it.

8. Carcinoma adheres quickly to the deep-seated parts, especially the pectoralis muscle.

9. Involves the lymphatic system in the early stage of the disease, which is always steadily progressive.

10. The morbid growth will surely and speedily return; usually in some other part of the body and a fatal termination rapidly supervenes.

11. The general health is quickly impaired, the cachexia becoming very marked in the early stage of the disease.

12. The progress of the disease is attended with severe pain.

Having pointed out the most essential differences between sarcoma and carcinoma of the breast, I shall now consider briefly, the chief characteristics of the former disease in this organ. It should, however, be remembered that while speaking of the morbid growths in the mammary gland, I do not confine myself to the spindle-celled sarcoma, but also include all the varieties of sarcomatous growths found in this organ. I have taken this liberty in order that I might be enabled to draw a dividing line between sarcoma and carcinoma.

The sarcomatous growths found in the breast were designated by Rindfleisch by the following names:

cysto-sarcoma fibrosum, cysto-sarcoma mucosum, sarcoma peri-canaliculare, sarcoma pericanaliculare diffusum. The sarcoma which is developed in the mammary gland is accompanied by a proliferation of cells of the lactiferous ducts. Cornil and Ranvier, who have studied very carefully these morbid growths have designated two forms, one in which the sarcomatous tissue is developed in the lobules at those points where the lactiferous ducts are the most numerously distributed, or, as is thought by some, within these minute canals.

In the other form of this disease, the granulating sarcomatous tissue which is supposed to originate within the lactiferous canals, pushes back their walls, extends into them in the form of simple papillæ, which distend these tubes and frequently become attached to them by a narrow pedicle. This growth is distinguished from the former by a more rapid development of the tumor and also by a more luxuriant proliferation of the foliated and papillar excrescences into the lactiferous ducts.

This condition becomes more marked when, besides the distention and distortion of the lactiferous ducts, there is also a collection of mucoid secretion within them, which gives rise to irregular sacs in which fluctuation may be detected. These sacs are the lacunary cysts which are so frequently mentioned by the various authors in their writings on sarcomatous tumors of the breast. Billroth has described this variety under the name of adenoid sarcoma, and many other authors have classified it with adenoid tumors, thus accounting for the fact that many morbid growths, so-called adenoids, have returned after their removal by operation.

The symptoms of the spindle-celled sarcoma differ somewhat from those which have been described in the above. This morbid growth, when it makes its appearance in the breast, is commonly indicated by roundish nodules, which have their origin in the

connective tissue surrounding the lactiferous ducts.

The pathological and microscopical appearances of the spindle-celled sarcoma of the breast is very similar to those presented by the same morbid growth in other parts of the body. Rindfleisch's¹ sarcoma *peri-canalicularare diffusum* is "an entirely uniform, yet mostly spindle-celled sarcomatous mass, within which the lactiferous ducts are distorted and distended into widely gaping clefts. This tumor also begins in the surroundings of the ducts, thence, however, passes over the entire interstitial tissues." The spindle-celled sarcoma of the skin, where this neoplasm is common, frequently shows itself in the form of a small tumor, a mere excrescence, which soon involves the entire thickness of the integument, and its breadth may exceed its thickness. This growth sometimes ulcerates at an early day, presenting a raw surface, covered with granulations which readily bleed, and soon become covered with a scab which falls off and is renewed from time to time. There is a form of spindle-celled sarcoma which takes its origin in the connective tissue beneath the skin, possessing many of the characteristics of the encephaloid sarcoma, although its consistence is more decidedly firm, occasionally even hard, where one observes fewer blue veins zig-zagging under the skin on the surface of the tumor and about its periphery. The skin, in some of these cases, remains a long time intact, when the disease is primary in its character, much longer than in cases of encephaloid sarcoma. The tumor, likewise, possesses a marked degree of mobility, but should this morbid growth happen to be attached to the aponeurosis it will then become almost immovable. Therefore, when a spindle-celled sarcoma originates in the periosteum it is naturally immovable. In these cases there is felt a mass which seems to be incorporated with the bone, even though

¹ A Text-book of Pathological Histology, p. 595, Trans. Phila., 1872.

only a lateral projection from it, while in other cases it envelops the whole surface of the bone like a cushion.

These tumors are commonly attended with little or no pain. They also develop very slowly, in fact their action is more torpid than that of the spindle-celled sarcoma of the soft parts. It is likewise thought that this tumor, when it has its origin in the periosteum is much more fatal than when it is confined to the soft tissues, a result which may, *in some measure be due* to the fact that owing to the severe character of the operation required to remove the morbid growth it is commonly postponed so long as to fail in accomplishing the object sought. The constitutional infection in the case of any spindle-celled sarcoma unquestionably takes place through the circulatory system, the blood carrying with it the contaminating juices of the tumor while the lymphatic vessels take but little or no part in this morbid process. It therefore follows that there is seldom found an enlargement of the lymphatic glands during any stage of its progress.

The spindle-celled sarcoma destroys life by a contamination of the general system, or by its pressure on important organs. When a spindle-celled tumor is removed, it may without doubt be reproduced, in fact this is the usual course which this disease takes; but in some patients the health remains good in spite of a multiplicity of relapses. Birkett has cited a case in which sixteen operations were performed within nine years, and where the general health remained unimpaired. A fact worthy of mention, but one that is also observed in other kinds of tumors, is the unfavorable action arising from a traumatism involving morbid growths, in which it is not unfrequently the case that an essentially harmless and chronic disease may take on a very rapid and unfavorable action in consequence of a severe blow or concussion. In connection with this statement it is not difficult to comprehend the effect produced by incomplete operations on various tumors, which, by

these operations, are caused to take on a high state of activity, frequently with a fatal result; thus a harmless fibro-polypoid growth situated in the nasopharyngeal cavity, frequently becomes sarcomatous following an incomplete operation for the removal of the primary tumor.

When the spindle-celled sarcoma is reproduced after its removal the secondary tumor may consist of essentially the same texture as the primitive; but there is a marked tendency in these growths to become more and more embryonic in their nature, thus precipitating the progress of the malady. The practical lesson taught by these observations is the avoidance of incomplete operation.

The ganglionic engorgement, which is rarely observed in cases of spindle-celled sarcoma, is merely irritative, and consequently entirely disappears after the removal of the morbid growth. The absence of this ganglionic engorgement is a peculiar characteristic and constitutes a marked difference between the encephaloid and spindle-celled sarcoma, ganglionic infection being rather common in the former, but exceptional in the latter.

It must be confessed that our knowledge of the etiology of the spindle-celled sarcoma is very limited and unsatisfactory. If we can believe statistics, the spindle-celled sarcoma of the soft parts is much more common among women than among men, the proportion being about two to one; but the order of frequency of occurrence in the sexes is reversed when this disease attacks the long bones. This disease occurs in all classes and at all ages, but most frequently in the middle period of life. It is very rare in children; nevertheless a case has been reported by Schwartz in which the disease attacked the tibia in a young infant. There are also reported cases of well-marked spindle-celled sarcoma of the skin which were discovered at birth, and it is also believed that the existence of excrescences or slight defects in

the integument present weak points which predispose to this degeneration. In addition to these etiological conditions, there should be mentioned the effects of repeated or continuous irritations, bruises, or contusions which have a definite action in the origin and development of sarcomatous growths, especially the spindle-celled sarcoma. There are innumerable cases cited in support of the views expressed in the preceding sentence, and it is impossible to deny their importance. It has likewise been clearly shown, that, if a sarcoma which already exists, be subjected to a traumatism, this action may become the cause of much greater activity in the morbid process. Inflammation, suppuration, and other unfortunate complications may now arise in rapid succession, hastening the disease to a fatal termination.

The diagnosis of spindle-celled sarcoma cannot be based on any pathognomonic signs. It is by carefully weighing the symptoms as a whole that we are able to diagnosticate the malady. Even a very careful examination of all the symptoms, in many cases, does little more than supply a serious presumption in favor of its nature, without enabling the surgeon to say positively that the tumor before him is a true spindle-celled sarcoma. Thus when this tumor it developed within an aponeurosis *e. g.*, in the fascia lata or some other fibrous tissue, it is rendered immovable, a fact which, when considered in connection with our knowledge of the nature of this neoplasm establishes a presumption in favor of its character. The same, or even greater difficulty, may be experienced in establishing a correct diagnosis in cases of spindle-celled sarcoma when it takes its origin from the periosteum, where it may be readily mistaken for an exostosis, whilst in other localities the question of an enchondroma may lead to a similar difficulty. In certain organs, the breast for example, the spindle-celled sarcoma may be mistaken for a scirrhus, carcinoma, or even an encephalo-

loid cancer. However, it should be remembered that the spindle-celled sarcoma develops more slowly, that it maintains a more independent existence, does not attach itself so firmly to the surrounding tissues, the skin which covers the morbid growth remains a long time intact; finally, enlargement of the glands in the axilla never takes place in the early stage of the disease, and very rarely even in the late. A small sarcoma in the skin may be readily mistaken for an epithelioma, but, if the subject is young, the age of the subject should be regarded as presumptive evidence in favor of the sarcomatous nature of the growth. It would seem that there should be very little trouble experienced by a surgeon in diagnosing a secondary sarcomatous growth appearing in a cicatrix following the removal of the primary disease; but even this may at times be very embarrassing. It is well known that a cicatrix may become reddened and thickened at various points, showing hard excrescences which are properly designated as cicatricial keloids, and that these are commonly regarded as inflammatory products. This peculiar growth frequently marks the former site of every suture employed in the treatment of the wound, or more accurately stated, these keloids form on every suture cicatrix. It may therefore become difficult, in some rare cases, to decide whether the morbid development appearing on a cicatrix is really a sarcomatous growth or not, but the surgeon need not long remain in doubt in these cases. The spindle-celled sarcoma will continue to grow steadily, but the keloid will soon come to a perfect halt and finally shrink and shrivel up.

The prognosis of spindle-celled sarcoma, is without doubt, very grave. Abandoned to itself it advances steadily, and ultimately leads to a fatal termination, either by the general contamination of the system, or local organic disturbance of important vital functions. Furthermore, the fact should not be lost sight of that the spindle-celled sarcoma soon re-

turns after its removal, that it may ultimately become constitutional, and likewise multiply in various organs of the body; notwithstanding all this, the prognosis is much less grave than in cases of carcinoma, or even of encephaloid sarcoma, its progress being slower and the surrounding tissues much less rapidly involved.

It is undeniable that the well-being of the patient is served by an early operation. A considerable number of patients on whom operations have been done under favorable circumstances have remained perfectly well for many years and finally died from other causes. The prognosis, whether favorable or otherwise, must depend largely on the locality of the tumor and the performance of an early or late operation—the former giving the patient the better chance for recovery and long life—while the complete removal of the morbid growth gives the highest degree of safety attainable in these cases; therefore any obstacle in the way of the full accomplishment of this object must influence, unfavorably, the opinion of the surgeon in the question of prognosis.

The treatment of the spindle-celled sarcoma is necessarily entirely surgical; and the earlier the operation is performed the better will be the chances of the patient. It should be carefully remembered that every surgical operation, when possible in these cases, ought to be made before there is involvement of the lymphatic glands or impairment of the general health. Another very important rule in the management of these growths is, that *every particle of the tumor should be removed; let not a single morbid cell remain behind*; if a long bone is involved, amputation is the only effectual remedy. This operation should be performed eight or nine centimetres above the apparent limit of the disease, as shown in the bone substance—better still, where practicable, to amputate through an articulation, above the morbid growth, because there are frequently found in the medullary canal sar-

comatous cells at a much higher point than in the bone. The performance of the complete operation is recommended by the following observations: First, ganglionic enlargement, when it exists, is promptly relieved by this procedure; secondarily, the contaminating juices stored up in the morbid growth, as well as the danger arising from its pressure on important organs, is completely removed, and the development of constitutional symptoms is postponed. Incomplete operations should not generally be performed; since this procedure accelerates the development of the morbid growth and gives it, in many instances, a malignant character; but there are unquestionably some cases in which the incomplete operation is absolutely necessary and consequently justifiable, thus, impending suffocation caused by a sarcomatous tumor would justify this procedure.

In connection with this paper I desire to report a case of spindle-celled sarcoma, and, likewise to present the pathological specimen, which, I think, will be accepted as typical of the disease; but my chief interest centres in the complications rather than in the tumor itself.

C. F. H., school boy, æt. 12, born of healthy parents, nervous temperament, somewhat delicate, accustomed to complain of headache while in daily attendance at school, fond of athletic sports but rather easily fatigued, had passed safely the diseases of childhood; health equally as good after an attack of scarlatina, occurring when the child was about 9 years old, as it had been previously. The tumor was discovered by the mother about November 1, 1885. It was then about the size of a goose egg, and was situated posterior to the middle third of the left femur. The patient, notwithstanding the dimensions to which the growth had already attained, had never complained of any pain or other inconvenience that could be attributed to the tumor. The family physician was finally called and continued in charge of

the case until the patient died, which occurred Jan. 5, 1886. In this case there was observed, about the 1st of December, a mottled appearance of the integument, covering some parts of the tumor, which at first consisted of a few isolated red spots. These continued to increase in number and also to enlarge their dimensions. Soon there was observed a change in their color—the spots which primarily were red, slowly turning to a bluish-brown, then to a brownish-black; and inasmuch as new spots were continually making their appearance the parts presented a variegated and mottled aspect. These changes in the skin took place without any apparent elevation of the spots above the surrounding integument. I saw the patient in consultation with the attending physician, about the 20th of December. The tumor was then about four-fifths as large as when I removed it, January 4. The entire surface of the integument covering the morbid growth was mottled as already described. The tumor occupied at this time about the same relative position on the thigh as when first discovered by the mother, although it had increased considerably in size.

I did not see the patient again until the day that the operation was performed. The tumor, in the meantime had changed in form (being less oval than it was), had increased in size, and had descended—the upper border being one inch lower than it was when I made my first examination, and the mottled surface of the integument was now three times greater than at my first visit. The superficial veins covering the tumor were not much enlarged. The patient was anæsthetized—a linear incision was made of sufficient length to enable me to remove the entire morbid growth which was found imbedded in the deep connective tissue—its anterior surface approaching closely to the periosteum of the femur, the sciatic nerve passing through one third of its lower portion, grooving deeply the upper two-thirds of posterior surface.

The operation was speedily accomplished and attended with very little loss of blood. There was some traction made on the sciatic nerve while liberating it from the tumor; it was also slightly incised at one point, and was completely detached from all the surrounding tissues throughout the entire length of the tumor.

The operation was performed under strict antiseptic precautions—the nerve was placed within the wound—drainage tubes introduced and the wound closed with iron wire sutures. The shock caused by the operative procedure was apparently slight and the patient reacted very well; but within a few hours it was observed that the mottled appearance which has been previously mentioned was extending very rapidly over the limb; and within thirty hours after the completion of the operation the gangrene had reached the toes and abdomen. The patient survived the operation about thirty-four hours.

I sent the tumor for examination to Dr. Frank Ferguson, pathologist of the New York Hospital, who reports as follows: "My Dear Dr. Watson: I send you the following statement of the tumor received from your office on Saturday last. The tumor measures, vertically, 12 centimetres; laterally, 8; and in its antero-posterior measurement, 6 centimetres. It weighs 304 grammes. Tumor is lobulated, and grooved in the median line posteriorly, to the depth $2\frac{1}{2}$ centimetres, where it received the trunk of the sciatic nerve.

"It is limited by a delicate fibrous capsule except at the point of attachment of muscles. In these locations the tumor is directly continuous with the fibrous tissue of the muscles. It is composed of numerous small spindle-cells and much fibrous tissue. The vascular supply is abundant and the vessel walls are composed of tumor tissue. Diagnosis: Fibrosarcoma. Yours sincerely, Frank Ferguson."

The chief interest in this case attaches to the in-

volvement of the sciatic nerve in the morbid growth which unquestionably caused the vaso-motor disturbances and ultimately the gangrene following the operation.

